

**Condition: Bernard-Soulier syndrome**

**Pathogenesis:** Congenital (autosomal recessive)

**Affliction:** Adhesion to von Willebrand Factor (primary haemostasis)

**Deficiency:** Glycoprotein Ib-IX-V complex

**Treatment:** NovoSeven (factor VIIa recombinant) preoperative (2 hour half life)

**Local Haemostatic Measures:**

- Pressure
- Tranexamic acid
- Local anaesthetic (epinephrine)
- Sutures (lacerations)
- Bipolar
- Laser Diode
- Silver nitrate

**Patient: Female, 62**

**Diagnosis: LL8 pulp necrosis, symptomatic apical periodontitis**

**Treatment:**

- RCT UL8

**Key Issues/Points:**

- Very high bleeding risk with severe Bernard-Soulier, decision to avoid XLA wherever possible
- Tranexamic acid and recombinant factor VII prior to treatment
- Use of buccal infiltration with 4% articaine with 1:100,000 epinephrine only, ID block and lingual infiltration contraindicated
- Short half life of NovoSeven adds complexity to treatment
- Post operative haematology review

Plan

1. Oral Tranexamic acid 1g TDS for 5 days starting the day before
2. Novoseven 6mg 30-60 minutes pre procedure.
3. HLA matched Platelets on standby.
4. Post dental review

**Left – Haematology plan**

**Below – Treatment photographs and radiograph**

